Tessier 30 with sublingual dermoid

Mukherji Srijon*1, Dey Annuradha2, Suba Santanu3

1Chief Consultant, Department of Oral and Maxillofacial Surgery, Calcutta Institute of Maxillofacial Surgery and Research, 200, Rajdanga Nabapally, Kolkata-700107, West Bengal, India.
2Junior Consultant, Department of Oral and Maxillofacial Surgery, Calcutta Institute of Maxillofacial Surgery and Research, 200, Rajdanga Nabapally, Kolkata-700107, West Bengal, India.
3Assistant Professor, Department of Plastic Surgery, Medical College, Kolkata-700073, West Bengal, India.

ABSTRACT

Tessier 30, also known as median midline cleft of the lower jaw, is a rare congenital anomaly presenting as cleft in the lower jaw that includes the tongue, the mandible, lower lip and the chin. There are less than 80 reported cases in the English medical literature since its discovery in 1819. We present one such case of Tessier 30 with presentation of cleft in chin and mandible along with ankyloglossia and a sublingual dermoid cyst that we reconstructed. 2 years post-operative follow up has been shown to be satisfactory.

Keywords: Dermoid; facial cleft; Mandibular cleft; Tessier 30; Tessier cleft.

INTRODUCTION

Tessier 30 cleft or “median midline cleft”[1] was first termed by Dr Paul Tessier in 1976 but the features were first reported by Couronne in 1819. (Figure 1) Tessier 30 in the minor forms just affect the soft tissue of the lower lip but may extend to involve mandible, tongue, uvula, strap muscles of the neck, thyroid cartilage, clavicle and sternum. [2,3] Dermoids in the midline of neck have also been reported. [2] We present a case of Tessier 30 involving the chin, mandible, tongue along with a sublingual dermoid cyst between the mandibular processes. In this case we also show an interesting use of the cyst epithelium for intraoral reconstruction.

Case History:

A 3 year old boy presented with cleft of lower lip leading to incomplete lip seal during swallowing and tongue attached to floor of mouth. The clinical examination depicted midline cleft of the lower lip. Fusion at the mandibular symphysis was absent, leading to two mobile pieces with a diastema between the 7A, 8A and ankyloglossia. (Figure 2, Figure 3) Other systemic anomalies were ruled out. Surgical correction was planned under general anesthesia for bony and soft tissue repair in a single stage.
The scar tissue around the cleft was marked and excised (Figure 4). Upon dissection, a sublingual dermoid cyst was found extending between the two mandibular segments. (Figure 5)

Release of ankyloglossia was performed. (Figure 5) Usually, a diamond shaped raw area; half on the ventral surface of tongue and half on the floor of the mouth were created. Usually, we medialize the edges for closure which post operatively result in scar contracture and loss of tongue freedom to some extent.

Here, traditional linear closure of the ventral aspect of tongue was performed. It was evident that after excision of scarred cleft tissue and release of ankyloglossia, the remaining soft tissue was inadequate. (Figure 6) The cyst fluid was drained and epithelium was spread inside out and used to augment the floor of the mouth, alveolar mucosa on the labial side, labial sulcus. (Figure 7) It mucosalized fast. (Figure 8)

The mandibular cleft was explored with minimal periosteal stripping. Segments were aligned at lower border and fixed in reasonable occlusion with trans-
Figure e-soft tissue in cleft region therefore requires a pair to 10 years of age to be performed. W.B. Saunders; 1990.

Repair of implant requiring secondary surgery is performed in adults, further orthodontic and orthognathic correction becomes essential. A series of soft tissue refining surgeries are required in cases of Tessier 30 cleft extending to the neck.

2 year post operatively our case shows no significant scar contracture or indicates need of revision. We used transosseous wiring for the mandibular processes as it was cost effective, preserved the developing tooth buds and spared a secondary implant removal procedure. No major retrusion of mandible noted in postoperative period as we fixed the mandible at early age before the advent of major growth spunts. One of the objectives to achieve in these surgeries is to achieve adequate movement of the tongue. The freedom of tongue achieved at end of 2 years was also significant. (Figure 9)

DISCUSSION

Reported by Couronne in 1918, “median midline cleft” of lower lip was first termed as Tessier 30 in 1976. The clinical presentations may be just soft tissue involvement of lower lip or extend further as cleft in mandible and/or ankyloglossia with/without a bifid uvula along with midline dermoids. Often thyroid cartilage show underdevelopment. Widely spaced clavicles and bifid manubrium sterni may be associated with Tessier 30. Presence of preternal tags and ventriculo-septal defect in these patients are also documented. Oostrom et al 1996, classified the Tessier 30 cases into soft tissue involvements, hard tissue involvements and associated anomalies depending on structures involved.

First branchial arch develops in the 7th week of intrauterine life which further divides into the right and left mandibular processes that fuse in the midline to form the mandible. Severe hypoplasia of the mandibular process may give rise to the midline cleft or mesenchymal cells may invaginate between the mandibular processes and prevent forming of complete mandible. Till 2015 only 80 such cases have been reported in the English medical literature- stating it as a rare congenital anomaly. Often thyroid cartilage show underdevelopment. Widely spaced clavicles and bifid manubrium sterni may be associated with Tessier 30. Presence of preternal tags and ventriculo-septal defect in these patients are also documented. Oostrom et al 1996, classified the Tessier 30 cases into soft tissue involvements, hard tissue involvements and associated anomalies depending on structures involved.

First branchial arch develops in the 7th week of intrauterine life which further divides into the right and left mandibular processes that fuse in the midline to form the mandible. Severe hypoplasia of the mandibular process may give rise to the midline cleft or mesenchymal cells may invaginate between the mandibular processes and prevent forming of complete mandible. Till 2015 only 80 such cases have been reported in the English medical literature- stating it as a rare congenital anomaly.

The timing for surgery should be as early as possible to prevent hypermobility of segments, respiratory and feeding difficulty but Armstrong, 1996, deferred the mandibular repair to 10 years of age to prevent damage to the developing permanent tooth buds. Minor soft tissue cleft were corrected with “V” excision of cleft followed by a Z-plasty yielding a comparatively esthetic result as scar contracture is less.

The surgical intervention is either single staged or multiple. In infants, the soft tissue closure performed first to help the patient form a complete lip seal for swallowing. The ankyloglossia may be corrected at a later stage while performing the hard tissue repair. Titanium plates fixation are preferred but require a removal procedure as in the symphysis region they may consequently retard the growth of the mandible. Rao A.N.V, 2015, therefore emphasized on use of bioresorbable plates. Hard tissue repair often requires iliac bone graft or rib graft. When primary repair is performed in adults, further orthodontic and orthognathic correction becomes essential.

Patient had an uneventful postoperative healing phase and achieved lip seal competence and showed free movement of tongue. The patient was on subsequent review cycle for 2 years. (Figure 11, Figure 12, Figure 13)

REFERENCES


5. Rao A.N.V, Complete Midline Cleft of Lower Lip, Mandible, Tongue, Floor of Mouth with Neck Contracture: A Case Report and Review of Literature, Craniomaxillofacial Trauma and Reconstruction Vol. 8 No. 4/2015
7. Ali AAA, Tessier Number 30 Median Mandibular Cleft With Congenital Heart Anomalies in Qena, Egypt, The Cleft Palate-Craniofacial Journal 56(2): May 2018